

# Congenital atrichia associated with nevus flammeus: A rare association

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## ABSTRACT

Congenital atrichia is the absence of hair over the entire body at birth, an inherited condition that may be isolated or associated with other anomalies. Herein we report a case of isolated congenital atrichia with nevus flammeus.

**Key words:** Congenital atrichia, infancy, nevus flammeus

## INTRODUCTION

Congenital atrichia is the total absence of hair from birth, usually an inherited condition. It may occur as isolated<sup>[1]</sup> or associated with other anomalies<sup>[2]</sup>. Congenital atrichia may be associated with papular lesions on the face, neck, trunk, or limbs<sup>[3]</sup> that may not be present at birth and may occur later in life. These are also a few syndromes associated with congenital alopecia such as Moynahan syndrome, hidrotic ectodermal dysplasia and progeria.

## CASE REPORT

A six-month-old baby born of consanguineous parentage (second-degree relatives), was brought to the outpatient department for scanty growth of scalp hair for the past three months. A few tufts of scanty hair were present on the scalp at the time of birth. There was complete absence of hair on the eyebrows and eyelashes. There was no history of hypertension, atopy, loss of hair, or any autoimmune disorder among the family members. The baby was otherwise healthy for the past six months. Developmental milestones and vaccination schedule were normal. Sweating and body temperature was normal. There was no history of epilepsy. Her elder sibling (two years old) was also normal.

Examination revealed a sixmonth-old baby with complete loss of hair on the occipital and both temporal regions of the scalp [Figures 1 and 2]. Partial loss of hair was present on the parietal and frontal areas of the scalp with the present hair being terminal [Figure 3]. There was complete

absence of superciliary and ciliary hair and absence of hair elsewhere on the body [Figure 4]. Hair pull test was negative. There was no visible inflammation at the follicular orifices, hypopigmented streaks suggestive of alopecia areata were not seen. Mucosae and nails were normal. An asymptomatic erythematous-to-pink blanchable patch with a well-defined border of 3 × 4 cm on the occipital area was present.

On trichoscopy, the hair present on the scalp was terminal with moderate thickness and no perifollicular pigmentation, inflammation, or occlusion [Figure 5]. On microscopic examination, the hair shaft and hair bulb were normal. Vitamin D3, serum calcium, and immunoglobulin E levels were normal. Biopsy was not done as the parents did not consent to it.

## DISCUSSION

Congenital atrichia can present as total loss of hair at birth or with scanty hair on scalp, which may later shed within a few months after birth. It may be inherited as an autosomal dominant or autosomal recessive disorder.

Vitamin D-dependent rickets type II A associated with alopecia and alopecia universalis should also be considered. There are a few syndromes associated with congenital alopecia such as Moynahan syndrome, hidrotic ectodermal dysplasia and progeria [Table 1].<sup>[2]</sup> There have been several reports of congenital atrichia,<sup>[3,4]</sup> though a few of them are isolated and not inherited.<sup>[1]</sup> There have also been case reports of

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**Figures 1:** Temporal and occipital regions with complete loss of hair and occipital area with erythematous blanchable patch



**Figures 2:** Temporal and occipital regions with complete loss of hair and occipital area with erythematous blanchable patch



**Figure 3:** Partial loss of hair on parietal and frontal regions

congenital alopecia areata in children of two to five years of age.<sup>[5]</sup> A solitary case of congenital atrichia has been reported with situs inversus and mesocardia.<sup>[4]</sup> Congenital atrichia may be confused with congenital alopecia universalis, but the distinct features such as a history of consanguinity, loss of hair either on eyebrows or eyelids and scant body hair and vitamin D deficiency in the form of rickets may suggest congenital atrichia. The presence of an asymptomatic erythematous-to-pink blanchable patch with well-defined borders on occipital area in our case was diagnosed as nevus flammeus as it was present since birth.

In our case, the differential diagnoses were narrowed down to congenital atrichia and congenital alopecia areata progressing

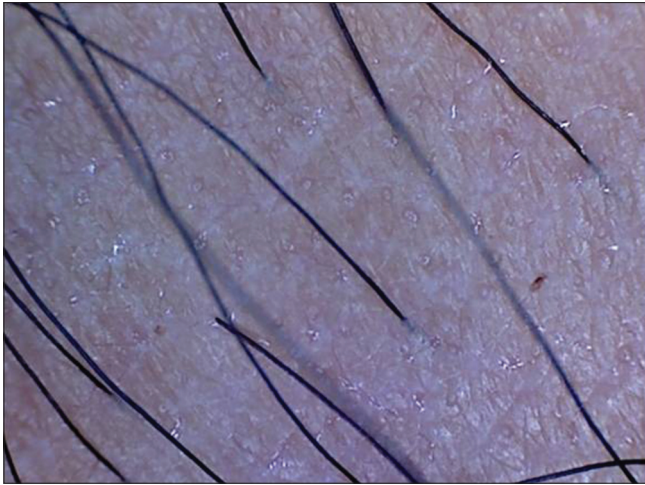


**Figure 4:** Complete loss of hair on eyebrows and eyelids

**Table 1: Various case reports and reference numbers**

Clinical condition	Case reports	References
Moynahan syndrome		[7]
Ectodermal dysplasia		
Progeria		
Papular lesions	Bansal <i>et al.</i>	[3]
Situs inversus and mesocardia	Sacchidanand <i>et al.</i>	[4]
Hereditary simple hypotrichosis	Ferrando <i>et al.</i>	[8]
Palmoplantar hyperkeratosis + mental retardation + early loss of teeth		[9]
Nevus flammeus	Present case	-

towards universalis. There was no history of epilepsy and hyperhidrosis, and levels of vitamin D3, serum calcium, IgE are normal. Alopecia areata is common in children, though it



**Figure 5:** Trichoscopic image with terminal hair and patent open follicles with no hair

is rarely reported in the neonatal period or in infancy. Alopecia areata progressing to alopecia universalis in the neonatal period has also to be considered and has not been reported so far.<sup>[2,6]</sup> As the absence of hair on the eyebrows and eyelids at the time of birth rules out the alopecia areata, with the history of consanguinity in parents and absence of family history, the diagnosis may be concluded as an "isolated case of congenital atrichia associated with nevus flammeus."

The case is being reported as congenital atrichia has not been reported in an infant of six months. The association with nevus flammeus, a sporadic form, and the trichoscopic picture

is reported for the first time in congenital atrichia. The patient is being followed up for any developments such as papular lesions on the face and body or for any epileptic episodes and mental retardation.

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